CASE REPORT

Ameloblastic Carcinoma: A Rare Malignant Tumour in Maxillofacial Region

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Abstract Ameloblastic carcinoma (AC) is a rare malignant odontogenic tumor with poor prognosis. It has an aggressive clinical course with extensive local destruction. It occurs primarily in the mandible. It may clinically present as a cystic lesion with benign clinical feature or as a large mass with ulceration, significant bone resorption and mobility of teeth in the affected region. Reliable evidence of its biological activity along with extensive local destruction, direct extension of tumor, lymph node involvement and metastasis to various sites (frequently lung) have been reported. Wide local excision is the treatment of choice along with regional lymph node dissection. Because of recurrence close periodic assessment of the patient is advocated. The authors discussed a rare case of AC of mandible; with metastasis to regional lymph nodes in a 45 year old male along with a long-term follow up.

Keywords Ameloblastoma · Ameloblastic carcinoma · Odontogenic tumour

Introduction

Ameloblastic carcinoma (AC) is a rare tumor which is characteristically differentiated from ameloblastoma by its highly aggressive nature and histological representation of malignancy [1]. There are two types—primary (which arises de-novo); and secondary (which is less common and arises from an existing ameloblastoma) [2]. However; the clinical features of these two types do not differ [3–6]. AC presents aggressive clinical features like rapid growth, perforation of the cortex and painful swelling [7, 8]. It may metastasize to regional lymph nodes or lungs. In rare cases, brain and multiple bone metastasis is reported [8]. This clinical case report describes the treatment modality for a 45 year old male patient diagnosed with AC which showed metastasis to the regional lymph nodes.

Case Report

A 45 year old male patient reported to the Department of Oral and Maxillofacial Surgery of with the chief complaints of mild pain and intraoral swelling in the left side of the jaw since past 6 months. The patient first experienced pain in the same region about 8 months back for which he consulted a dentist. Extraction of carious and grossly destructed mandibular left first molar was carried out at that time. A few days after extraction, patient noticed a swelling in that region which persisted along with mild pain till the time of his reporting to the Department.

Medical history of the patient was found to be non-contributory. Extra-oral examination revealed left sub-mandibular lymphadenopathy. On intra-oral examination, a diffuse swelling was observed in lower left vestibule, extending from 34 to 38 region. The overlying mucosa

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appeared normal in colour. Swelling was mildly tender on palpation, hard in consistency and had smooth ill-defined margins. The mandibular first molar had been removed completely. However root pieces of second molars were present (Fig. 1a, b). No clinical signs of trismus or paraesthesia were observed.

The panoramic radiograph showed an irregular, partially well-defined multilocular radiolucency in the mandibular body, extending mesio-laterally from lower left central incisor to the mesial aspect of third molar and supero-inferiorly from crestal region to the inferior border of the mandible. The locules within the radiolucency were more prominent in the distal aspect, whereas, mesially the radiolucency appeared largely to be having a single locule. Resorption of roots of the associated teeth and encroachment of the inferior alveolar nerve was also evident. The superior border of the mandible also appeared to have resorbed partially in the region (Fig. 1c). Computed tomography images confirmed the findings and showed an osteolytic lesion in the left mandibular body with erosion of both buccal and lingual cortical plates. The involvement of submandibular and cervical lymph nodes as well as the left submandibular salivary gland was also evident (Fig. 1d). The patient's chest radiograph was taken to rule out any lung lesion and it appeared to be normal.

A biopsy specimen was obtained under local anesthesia. Histopathological examination revealed connective tissue

stroma which was mature and fibrocellular. There were areas of odontogenic epithelium in the form of islands and follicles. Cord-like projections of epithelium in connective tissue were also observed. The follicles were lined by ameloblast-like cells with central areas showing stellate reticulum-like cells. The cells were highly pleomorphic with increased nucleus-cytoplasmic ratio, nuclear and cellular pleomorphism and areas of dyskeratosis. The stellate reticulum showed squamous cell metaplasia. One or two areas of necrosis were also seen. Some follicles showed areas of inductive effect with hard tissue formation (Fig. 2a, b). On the basis of histopathological investigations, a diagnosis of AC; secondary type (intra-osseous dedifferentiated) was made.

To rule out any distant metastasis, patient was later subjected to CT examination of the lungs and neck as well as an ultrasound of the abdomen and pelvis. There were no significant findings, except for enlarged submandibular lymphnodes.

During the surgery, an extensive surgical resection was carried out, which included left segmental mandibulectomy from left angle of the mandible to midline, taking a safe margin of 1.5 cm along with supra-omohyoid neck dissection. Immediate reconstruction was done with a stainless steel reconstruction plate, as the patient was not willing for further reconstructive procedure (Fig. 2c). Surgical specimens (Fig. 2d) were sent for histopathological examination

Fig. 1 a Preoperative photograph of the patient showing diffuse swelling in left side (arrow). b Panoramic radiograph showing multilocular radiolucent lesion in the body of the mandible. c Computed tomography images showing destructive lesion

Fig. 2 **a** Histopathology photographs (hematoxylin and eosin stain) showing follicles arranged with keratin pearls (original magnification, $\times 10$). **b** Histopathology photographs (hematoxylin and eosin stain) showing pre-ameloblast like cells which are highly dysplastic with

central cells showing squamous metaplasia (original magnification, $\times 40$). **c** Intra-operative photograph showing lesion after exposure and reconstruction with stainless steel reconstruction plate. **d** Surgical specimen

which again confirmed the histopathological diagnosis of AC; secondary type (intra-osseous dedifferentiated). Post-operative radio therapy was not advised.

Patient has been on a regular clinical follow-up regimen for the past 2 years, and has not presented any evidence of recurrence and/or metastasis in this duration (Fig. 3).

Discussion

Ameloblastic carcinoma (AC) is a rare neoplasm which poses a challenge in terms of its diagnosis, treatment and prognosis [7]. Similar to ameloblastoma, AC is also more commonly seen in the mandibular region than the maxilla [9]. Ratio between mandibular and maxillary occurrence of AC is reported to be 2:1 [9]. It has a wide age range of occurrence, with cases reported in patients between 10 and

84 years of age; with a mean age of 30.1 years [9–11]. No gender predilection is seen.

In the updated histological classification of the WHO in 2005, AC is classified as AC primary type; AC secondary type—dedifferentiated, intra-osseous; and AC secondary type—dedifferentiated, peripheral (carcinoma ex peripheral ameloblastoma) [3].

The clinical features of this tumor include swelling, pain, trismus, dysphonia, and mobility of associated teeth [9, 10]. As compared to ameloblastoma, AC presents with more aggressive clinical features like rapid growth, propensity to perforate cortex, pain and sensory disturbances [7].

In most of the cases, radiographic findings show an ill-defined radiolucency. However, focal radiopacity may also be detected within the radiolucent lesions [8].

As seen in our patient, AC may metastasize to the regional lymph nodes or lungs. In rare cases, brain or

Fig. 3 2 Year follow up photographs showing no evidence of recurrence

multiple bone metastases have been reported [8]. Avon et al. reported the need for adjunctive diagnostic examination after the diagnosis of AC, which includes a workup consisting of computed tomography, chest radiography and neck evaluation to determine the presence of nodal metastasis [9, 10].

The treatment of AC is controversial. Therapeutic guidelines do not clearly exist for AC because of its rarity [3]. Recommended surgical treatment usually requires jaw resection with 2–4 cm bony margin and consideration of contiguous neck dissection (both prophylactic and therapeutic) [11, 12]. A small number of reports have described radiotherapy and chemotherapy for the treatment of AC but the efficacy is not clear [3, 8]. Pre-surgical radiation is suggested to decrease the tumor size [11]. Most of the ACs are intraosseous; therefore, the effectiveness of radiation therapy must be considered critically. Use of gamma knife and stereotactic radiosurgery in the treatment of AC has also been reported [2].

ACs can recur locally 0.5–11 years after definitive therapy. Distant metastasis is usually fatal and may appear

as early as 4 months or as late as 12 years postoperatively. The common site for distant metastasis is lung. Distant metastasis can occur in the absence of local or regional recurrence [13].

Sufficient time should be given before reconstruction because of potential tumor recurrence [12].

Conclusion

While dealing with ameloblastoma its malignant nature should also be taken in consideration. Patients presenting with features of ameloblastoma should also be screened for its malignant form i.e. ameloblastic carcinoma. Although AC is a rare lesion, surgeon must be cognizant of its clinical and histological characteristics. It has an aggressive nature and is defined by malignant histopathologic changes. Hence, aggressive and complete removal of the tumor is recommended for successful treatment outcome. Because of recurrence and metastasis in regional lymph nodes and lung, meticulous long term follow up of such patients, monitoring of their health status and screening is also recommended.

Compliance with Ethical Standards

Conflict of interest Pushkar D. Gawande, Kiran Khande, Gaurav Agrawal and Amita Aditya declare that they have no conflict of interest.

Ethical Approval Approval of the institutional ethical committee was taken.

Informed Consent Informed consent was obtained from the patient.

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